Concomitant Conus Medullaris Arteriovenous Malformation and Sacral Dural Arteriovenous Fistula of the Filum Terminale

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Summary

A patient with a spinal intradural arteriovenous malformation (AVM) at the conus medullaris concomitant with a sacral dural arteriovenous fistula (AVF) of the filum terminale is reported. A 44-year-old-male presented with bilateral leg weakness and urinary incontinence for several months. Spinal angiography demonstrated two lesions: one was spinal intradural AVM at the conus medullaris supplied by the anterior spinal artery; the other was sacral dural AVF of the filum terminale supplied by the middle sacral artery.

Although multifocal spinal cord AVMs have been reported, this is the first case report of two different types of vascular malformations co-existing in one patient. The arterial supply of the dural AVF of the filum by the middle sacral artery is also first demonstrated in the literature. The patient was treated successfully by surgical approach for both lesions in the same operation.

Introduction

Spinal cord arteriovenous malformations (AVMs) are relatively rare¹, but they are important clinical entities because they may pro-

duce considerable morbidity. Their incidence is only about one-tenth that of brain AVMs. Recent advances in understanding of spinal vascular lesions have led to their classification into dural arteriovenous fistulas (AVFs) and intradural AVMs, which include juvenile and glomus AVMs and direct AVFs^{2,4}.

Dural AVFs produce symptoms of venous hypertension and cause congestion of the spinal cord, whereas intradural AVMs are usually high-flow lesions that elicit symptoms by hemorrhage or by ischemia from venous congestion of the spinal cord^{1,6}.

Multifocal spinal cord vascular malformations have been reported^{1,5}. Matsumaru et al⁵ reported 19 cases of multiple vascular malformations in 119 spinal cord AVMs. Most of the cases were metameric distribution or associations.

This case report describes concomitant spinal intradural AVM at the conus medullaris and a sacral dural AVF of the filum terminale. To our knowledge, this is the first case report of two different types of spinal vascular malformations co-existing in one patient.

The possible pathophysiology and the vascular malformations over the lumbosacral region and the filum, as well as treatment strategies, will be discussed.

Figure 1 Initial spinal angiography of selective left T11 intercostal artery shows a small intradural arteriovenous malformation (arrow) at the conus medullaris, which is located slightly to the right of the cord.

Case Report

A 44-year-old Danish male was well until several months previous to admission. He presented with progressive weakness of both legs and urinary incontinence. Initial myelography was performed in April 1996, revealing numerous serpentine vessels in the lumbar spinal thecal sac.

In May 1996, the first spinal angiography (figure 1) disclosed a small intradural AVM at the conus medullaris, which was supplied by the an-

terior spinal artery through the left T11 radiculomedullary artery. Relatively early venous drainage was observed down to the filum. The anterior spinal artery as well as the draining vein was tortuous and mildly dilated. Magnetic resonance imaging (MRI) was unavailable at the initial diagnosis.

The patient did not receive any treatment initially. However, the clinical symptoms progressed in the following couple months and he was transferred to our hospital for management.

The second angiography (figure 2) revealed the same findings as initial studies, a small AVM at the level of the conus medullaris with anterior spinal artery supplied via left T11 radiculomedullary artery. There was a small nidus or a direct arteriovenous shunt which was located slightly to the right of the cord. It was intradural spinal AVM superficial or around to the spinal cord.

The flow of this small AVM was not very high. The normal angiographic pattern of the basket of the conus medullaris was not well demonstrated. Descending venous drainage down to the filum was also identical to the first angiograms. Another unexpected arteriovenous shunt at the posterior margin of the sacral body at S2 level was found (figure 3). Selective middle sacral angiography revealed a dural AVF located at the midline at the level of S2 with ascending venous drainage. We believed this was a sacral dural AVF of the filum terminale.

The directions of these two distinct drainage veins were different. The superior one drained downward and the inferior one drained upward. Due to the small and tortuous features of the anterior spinal artery, the lesion of the spinal AVM could not be reached by endovascular access.

The patient had microsurgery performed on these two lesions in the same operation. He made a good recovery without any neurological symptoms. Three months later, the control spinal angiography (figure 4) including middle sacral artery showed no residual abnormal vessels.

The spinal AVM at the conus medullaris as well as the sacral dural AV fistulae of the filum were no longer visible. Follow-up MRI of the lower thoracic and lumbar spinal cord showed no evidence of spinal cord congestion.

Figure 2 Second angiography shows the spinal AVM (arrow head) at the conus medullaris with the same angioarchitecture as initial angiography. Arterial phase (A) shows the AVM supplied by the tortuous anterior spinal artery (arrow). Descending venous drainage (arrows) is noted in the venous phase (B).

Discussion

Spinal arteriovenous malformation (AVM) as well as spinal dural arteriovenous fistula (AVF) are relatively rare but important disease entities because they may produce considerable morbidity¹.

The major pathophysiological mechanism involved in spinal dural AVFs, regardless of their location, is venous hypertension. It was first proposed by Aminoff et al in 1974⁶, and the concept has been widely accepted. Acute neu-

rological deterioration may be caused by thrombosis of the draining veins.

The pathological presentations caused by spinal cord AVM are hemorrhages, including subarachnoid hemorrhage and hematomyelia. Venous hypertension and cord congestion also cause important neurological symptoms in spinal cord AVM. Only a large venous pouch can cause mass effect of the spinal cord^{1,7}. In Matsumaru's report⁵, the multiple spinal vascular malformations were not rare, particularly multifocal intradural malformations.

Figure 3 Selective middle sacral artery injection shows another sacral dural arteriovenous fistula. Lateral projection shows the sacral dural AVF with ascending drainage veins (A,B). Non-subtracted angiogram in lateral projection shows the exact level of the sacral dural AVF at S2 (C). Ascending venous drainage vein was observed in AP projection at the lumbosacral (D) and lumbar regions (E, F).

The series reported¹⁹ (16%) multiple vascular malformations in 119 spinal cord AVMs. Most of the cases were metameric associations or syndromes including Cobb syndrome, Klippel-Trenaunay-Weber syndrome and Parkes Weber syndrome⁸.

Unclassified associations also existed with bifocal intradural uni- or multimetameric lesions. The associated lesions were vertebral hemangiomas, cutaneous or peripheral vascular lesions, radicular AVMs, or even bifocal spinal cord AVMs. These kinds of lesions usually do not involve the dura⁵. They must be related to the absence of dural sinuses at spinal level.

In this particular case, the patient had two lesions, with one at the conus medullaris supplied by the anterior spinal artery and the other one at the posterior margin of the sacral body at S2 level supplied by the middle sacral artery. The upper one was obviously a small spinal cord AVM, although it might have been either a small nidus or a direct arteriovenous shunt.

The caudal one was not present in the first examination. It could have been either co-existing with the conus medullaris AVM at the same

period when the patient made the diagnosis or it might have developed after the AVM. It is possible to produce a secondary spinal dural AVF after hemodynamic change of the spinal cord caused by the cord AVM. There are several theories regarding development of the spinal dural AVF¹. But the venous pressure of the cord seems an important factor for the pathophysiology of spinal dural AVF. The directions of venous drainage of these two respective lesions were different.

They both drained through the tortuous medullary veins, but the upper lesion drained downward and the lower lesion drained upward. Such venous drainage may have worsened the venous hypertension of the spinal cord and progressed the neurological symptoms of the patient.

Considering vascular malformations at the lumbosacral level, there are three possibilities. The first is lumbosacral nerve root AVF or cauda equina AVM. The arteries of the shunt usually arise from dural arteries. Cases with supplying arteries from branches of the internal iliac artery and lateral sacral arteries have been

Figure 4 Control angiography 3 months later shows no residual abnormal vessels. Selective left T11 intercostal: A) arterial phase, B) venous phase and middle sacral artery, C) angiography in AP projections.

reported^{9,10}. The drainage veins are radicular veins and usually follow the nerve roots, which locate more laterally and near the region of the sacral foramen.

Few cases with intradural drainage upward through medullary veins were reported. Such kind of drainage direction might cause more significant clinical symptoms. The second vascular malformation at the lumbar level is AVM of the filum terminale^{1,11}.

The presented cases in the literature demonstrated that the filum terminale AVM had arterial blood supplied from the anterior spinal artery and drained into anterior or posterior medullary veins in ascending fashion. The third situation is dural AVF of the filum terminale. From the anatomic point of view, the filum terminale should be supplied by the anterior

spinal artery. However, the caudal ending of the filum terminale usually attaches on the dural at the level of S2. In this case, the dural AVF was fed by the small branches of the middle sacral artery at the level of S2. We believe this was a dural AVF of the filum terminale close to the dural attachment with the arterial supplied by the middle sacral artery, which was shunted into the filum vein and joined the medullary veins. A dural supply from the middle sacral artery has never been reported.

Spinal AVMs as well as spinal dural AVFs can be treated either by endovascular embolization or surgery. Early reports have described the use of particles for embolization, but more recent studies have revealed that long-term failure rates with this method are high¹².

Embolization using liquid polymers like n-

butyl-2-cyanoacrylate (NBCA) allows for more precise control with satisfactory results^{1,7,11}.

The treatment of patients who have multiple lesions is always difficult. The lesion responsible for the clinical symptoms has to be identified. The strategy should be decided on an individual basis as well as angioarchitecture of the lesions. In this particular case, the patient had a very tortuous and not very dilated anterior spinal artery. Embolization of the spinal AVM at the conus medullaris through the anterior spinal artery was very difficult. Besides, there was a second lesion, dural AVF of the filum terminale, with different feeding arteries. Both lesions had mildly dilated drainage veins with different fashions; both were possibly responsible for the clinical symptoms of the patient.

Embolization or surgical excision of any one of the lesions would not make sure that the venous hypertension of the spinal cord could be relieved. Surgical treatment for both lesions in the same operation was the best choice for this patient.

This case report highlights the possibility of two co-existing spinal vascular malformations, not only for different types of AVMs or AVFs but also for any locations of the spinal level, even though these vascular malformations may produce identical clinical symptoms. Treatment options should take both lesions into consideration including the types of the lesions, locations, angioarchitecture and clinical symptoms. Otherwise, we may not reach a satisfactory result if we leave either of the lesions untreated.

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